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Diabetes mellitus and cystic fibrosis

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Aims Children with cystic fibrosis may develop Type 1 diabetes mellitus or cystic fibrosis related diabetes mellitus. Good control of diabetes is known to be vital to preserve pulmonary function and growth. It is therefore important to determine an accurate blood sugar profile in order to construct an individualised insulin regimen.
Methods Children with fasting blood sugars greater than 7.1 mmol/l have an oral glucose tolerance test. If this reveals abnormal glucose tolerance then they have a 4 day continuous glucose profile utilising a Minimed glucose monitor. The profile is repeated once the child is on insulin or if there is continuing deterioration in pulmonary function or if steroids are introduced.
Results Four or even eight point blood sugar profiles do not always give sufficient information about blood sugar levels to construct an adequate insulin regimen for children with CFRD. The results indicate significant fluctuations in blood sugars especially with overnight feeds that are not well controlled on standard type 1 diabetes insulin regimens. We now have CF children on a wide variety of insulin regimens. Overnight feeds often require additional insulin, the addition of steroids requires a different regimen.
Conclusion Untreated or inadequately treated diabetes mellitus in children with CF can lead to reduction in growth and declining pulmonary function. The dietary requirements must continue so the insulin regimen must adapt to these requirements. We have demonstrated that individual children with CFRD and type 1 diabetes have very different blood glucose profiles that were not obvious on 4 or 8 point blood sugar estimations. The use of a 4 day continuous blood glucose profile has allowed us to individualise and monitor insulin regimens regardless of the dietary needs. We believe that this helps improve the overall health of the individual child.

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Spontaneous and reactive hypoglycemia in patients with Cystic Fibrosis

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Growth failure and diabetes (CFRD) are associated to anabolic defects in Cystic Fibrosis patients (CF). Insulin is choice therapy in CFRD and could be used to counteract nutritional decay and progression to CFRD in glucose intolerant patients (GI) but hypoglycemia could be a limiting factor. Postabsorptively many CFRD patients are normoglycemic and a subset of CF is spontaneously hypoglycemic.
Aims. To evaluate frequency of spontaneous hypoglycemia, and relate it to glucose tolerance and nutritional status
Methods. 118 CF without known CFRD received yearly OGTT (149 studies, 1.27 studies/patient) and nutritional status evaluation.
Results. 15.3% of CF (12.1% of studies) had postabsorptive glucose (PG) <60 mg/dl but 1 was GI and 1 was CFRD. In the whole series 18% of OGTT showed GI and 18% CFRD. Results in 15 hypoglycemic (40-57 mg/dl, HYPO) and 15 normoglycemic normotolerant (CON) sex and age pair-matched CF (16.6±1.2yr, range 11-24) were:

	Glucose (mg/dl)				Insulin (μU/ml)				HOMA	Weight	Height
	Bas	max	120'	AUC	bas	max	120'	AUC	IS	z-score	z-score
HYPO	51	180	113	10540	7	67	42	4550	5.3	-0.6	-0.5
	±1*	±11	±12	±1022§	±2	±10	±10	±614	±3.8	±0.3°	±0.4
CON	80	163	97	6555	7	64	39	3650	4.8	-0.1	-0.1
	±1	±10	±7	±826	±2	±11	±11	±598	±2.4	±0.3	±0.3

* p=0.04 and § p=0.005 (t-test). ° greater proportion of negative values, p=0.03 (χ^2 test)

The peak PG was similar to CON because of a marked increment in PG AUC. 7 HYPO and 4 CON had reactive hypoglycemia (<64mg/dl). Insulin responses and sensitivity were not different. More HYPO were underweight with a worse Shwachman score (58±3 vs 67±3, p=0.04).
Conclusions. Spontaneous hypoglycemia does not exclude GI or CFRD and is more frequently associated to nutritional status impairment suggesting an anabolic defect.

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Prevalence of osteoporosis and osteopenia in children and adolescents with cystic fibrosis

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The *aim* of our study was to determine the prevalence of osteoporosis and osteopenia in CF children and adolescents and to identify risk factors.
Methods: We studied 54 CF patients (23 male and 31 female) aged 4-21 years (mean age 12.5 yrs). Seventeen of them were in pre-pubertal and 37 were in mid- and post-pubertal stages of development. All patients suffered from pancreatic insufficiency and were taking 20 μg oral vitamin D daily. Serum 25(OH)D levels were within normal limits in all the patients (20.05±9.65 ng/ml), but were lower than those of healthy controls (39.64±8.94 ng/ml). None of the patients had been taking oral corticosteroids. Eighteen out of 54 patients had FEV_1< 50%. Forty-five of the 54 patients were chronically infected with Ps. aeruginosa. BMD was measured at the lumbar spine by dual energy x-ray absorptiometry.
Results: Twenty two of the 54 patients (41 %) had osteoporosis and 18 out of 54 patients (33%) had osteopenia. Osteoporosis was found in 41% of pre-pubertal and in 40.5% of mid- and post-pubertal CF patients. Osteopenia was found in 29% of pre-pubertal and in 35% of mid- and post-pubertal CF patients. In patients with FEV_1≥50%, osteoporosis was found in 25% and osteopenia in 42%. In patients with FEV_1<50%, osteoporosis was found in 72% and osteopenia in 17%. Chronic infection with Ps. aeruginosa was not found to be a risk factor for reduced BMD.
Conclusions: Osteopenia and osteoporosis are common findings in CF patients. Despite the adequate administration of vitamin D, serum 25(OH)D levels are lower in patients than in controls. Patients mostly at risk are those with reduced lung function.

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Bone mineral density in children with cystic fibrosis and the effect of vitamin D and calcium supplementation

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Introduction: Low bone mineral density (BMD) is a relatively frequent finding in individuals with cystic fibrosis (CF), particularly in adults. BMD might be related to vitamin D and calcium malabsorption, although the pathogenesis of low BMD in CF population is multifactorial.
The aims of this study were to ascertain BMD in children with CF and to evaluate the effect of vitamin D and calcium supplementation.
Methods: Anthropometry data, dual-energy X-ray absorptiometry (DEXA) BMD of lumbar spine (L1-L4), calcium urinary excretion (fU-Ca) were measured in 14 patients (aged 5.2–13.7 years, mean 10.0 years, 9 male, 5 female). Calcium and vitamin D supplementation were adjusted according to these data. The effect of treatment was evaluated after one year.
Results:

	Baseline			1 year after		
	min	max	mean	min	max	mean
BMD Z-score	-3.5	-0.7	-2.1	-2.5	0.2	-1.4
fU-Ca (mmol/24h)	0.53	1.31	1.00	0.31	4.94	2.76
Ergocalciferol supplementation (IU/day)	800	4000	3086	1600	8000	4400
Calcium supplementation (mg/day)		0		0	500	214

Conclusions: 1. Low BMD may be present in prepubertal CF patients despite of usually recommended vitamin D dosage. 2. Early detection of low BMD and consequent vitamin D plus calcium intervention may improve BMD. 3. Measurement of calcium urinary excretion is an advisable, non-invasive and cheap method for monitoring of vitamin D and calcium supplementation.